

PARENT FACT SHEET

Very long-chain acyl-CoA dehydrogenase deficiency (VLCAD)

CAUSE

VLCAD occurs when the very long chain acyl-CoA dehydrogenase enzyme is missing or not working properly. This enzyme's job is to break down certain fats from the food we eat into energy. It also breaks down fat already stored in the body.

IF NOT TREATED

VLCAD is variable and can cause mild effects in some people and more serious health problems in others. Symptoms may start in infancy or not until adulthood. There are three forms of VLCAD: Early, Childhood, and Adult.

TREATMENT OPTIONS

Your child will need to be under the care of a metabolic specialist and dietician. Treatment is usually needed throughout life.

- Your child needs to avoid going a long time without food. This is to prevent a rapid drop in blood sugar. These children should not go more than 4 to 6 hours without food and some may require more frequent feedings. It is important that these children be fed in the night – meaning you will need to be wake them up to eat if they do not wake up on their own – even if they are not hungry.
- Frequently, a low fat, high carbohydrate diet is recommended. Your dietician will help develop your child's food plan and no changes should be made without first getting the approval of the metabolic specialist and the dietician.
- Medium Chain Triglyceride oil (MCT oil) is often used as part of the child's food plan. You will require a prescription from the metabolic specialist to get this. The specialist and the dietician will assist you in how to properly use MCT oil.
- Some children take L-carnitine. It is a safe and natural substance that helps the body make energy. The metabolic specialist will decide if your child can benefit from this treatment. A prescription for this is also required.
- Contact your child's doctor immediately at the start of any illness. Children with VLCAD need to be treated in a hospital to prevent serious health problems.
- Avoid your child's prolonged exercise or exertion.

IF TREATED

Before diagnosis through newborn screening was possible, the early form of VLCAD was fatal. Now, with immediate and ongoing treatment, many infants are surviving. With prompt and careful treatment, people with childhood and adult forms of VLCAD can often live healthy lives with typical growth and development.

For more information go to the following website: <http://www.newbornscreening.info>